SHORT REPORT

Spontaneous closure of congenital coronary artery fistulas

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Abstract

Six cases of full spontaneous closure of congenital coronary artery fistulas, and one case of near closure, as seen by colour Doppler echocardiography, are presented. It is worth reconsidering the classical view that nearly all cases of spontaneous closure are eligible for surgical or percutaneous correction to prevent the development of significant and potentially fatal complications. As the natural course of coronary artery fistulas is still poorly defined, asymptomatic patients, especially those under 7 years old with small shunts, should be periodically followed up by echocardiography rather than be subjected to operative closure, even by catheterisation.

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Cases

The case records of seven children (four girls and three boys) with congenital CAF were reviewed. The baseline clinical and laboratory characteristics of these patients are shown in table 1. They came from three centres: Lille, France—patients 1 and 2; Leuven, Belgium—patients 3, 4, and 5; and Lyons, France—patients 6 and 7. All of the patients had been delivered without obstetrical problems.

The children were referred because of cardiac asymptomatic murmur. Their mean (SD) age at diagnosis in the outpatient clinic was 7.6 (5.7) months (range 2–18 months). In all of these normally developed children, a continuous murmur could be heard at the lower left sternal edge, in the fifth intercostal space in the midclavicular line or in the xyphoid area. The first and second heart sounds were normal. No patient had any symptom of cardiac failure.

ECGs and posteroanterior chest radiography, performed in all patients, were normal.

Two dimensional (cross sectional) and Doppler echocardiography, including colour flow mapping, showed the origin of the right coronary artery only was dilated in patients 2, 3, and 4, and the origin of the left main coronary artery only was dilated in patients 5, 6, and 7. In patient 1, the origins of both the left and right main coronary arteries were dilated. The diameter of the coronary artery origin exceeded 4 mm in all of the patients, and 5 mm in five of them (fig 1). The fistula drainage chamber was at the apex of the right ventricle in all patients, as indicated by continuous flow. One patient also had an additional congenital muscular ventricular septal defect (patient 4). The right ventricle was not dilated in any of the patients.

Congenital coronary artery fistulas (CAF), first described by Krause in 1865,¹ are characterised by normal aortic origin of the coronary artery involved but with a fistulous communication with the atria, ventricles or with the pulmonary artery, coronary sinus or superior vena cava. Such fistulas are the most common congenital coronary malformation. An increasing number of cases have been reported since the introduction of selective coronary arteriography and echocardiography.

Isolated cases of spontaneous or near closure of CAF have been described.^{2–14} We report six cases of full spontaneous closure and one case of near closure, as seen by colour Doppler echocardiography.

Table 1 Presentation, clinical findings, and outcome in seven patients with congenital coronary artery fistulas

Patient	Sex	Age at diagnosis (months)	Continuous murmur	Fistula course	QP:QS	Age at closure (years)
1	Female	3	2/6	RCA/LAD → RV	1	3
2	Female	7	2/6	RCA/LCX/LAD → RV	1.3	2
3	Female	11	3/6	RCA → RV	1	7
4	Female	9	3/6	RCA → RV	1	4
5	Male	18	3/6	LAD → RV	1	3.5
6	Male	2	3/6	LAD → RV	–	1.5
7	Male	3	2/6	LAD → RV	–	3

LAD, left anterior descending coronary artery; LCX, left circumflex coronary artery; RCA: right coronary artery; RV, right ventricle; QP:QS, pulmonary to systemic blood flow ratio; – no information available.

Figure 1 Two dimensional and Doppler echocardiography, including colour flow mapping, showing a > 5 mm diameter dilation of the left main coronary artery and left anterior descending coronary artery.

Figure 2 Selective angiography of the right coronary artery showing major dilation of the vessel, especially within its distal segment draining to the apex of the right ventricle.

The CAF diagnosis was based on coronary artery dilation associated with abnormal, high velocity flow at the apex of the right ventricle in patients with a precordial continuous murmur.

Cardiac catheterisation was performed in all but two cases (patients 6 and 7). Oxygen saturation data were normal in four patients. The pulmonary to systemic blood flow ratio (QP:QS) was 1.3 in patient 2. Intracardiac pressures were within normal limits in all patients. In three cases where dilation of the right coronary artery origin was observed by echocardiography (patients 2, 3, and 4), selective coronary angiography confirmed the isolated dilation of the vessel connected to the right ventricle by a fistula in two cases (fig 2) and an associated moderate dilation at the origin of the left anterior descending coronary artery in the third case. In three patients with a dilated origin of the left coronary artery (patients 5, 6, and 7), selective coronary angiography confirmed the dilation. It also revealed moderate dilation of the right coronary artery in one case (patient 5). In those three cases, the site of drainage was the apex of the right ventricle.

Clinical examination and echocardiographic follow up was undertaken for all patients. They all remained asymptomatic. No cardiac murmur was heard for 15–96 months (mean 40 (12) months) after the initial diagnosis of CAF.

Figure 3 Two dimensional echocardiography revealing the diameters of the left main coronary artery and left anterior descending coronary artery to be normal. Ao, aorta, PA, pulmonary artery.

Echocardiographic findings 1.5–7 years (mean 3.4 (1.8) years) after initial diagnosis were normal in patients 1–6, showing the size of the coronary artery origin to be normal with no abnormal flow into the right ventricle (fig 3).

In patient 7, the left coronary artery remained slightly dilated at two dimensional echocardiography mode. The Doppler mode revealed only a diastolic flow at the apex of the right ventricle because the CAF was nearly closed.

A total 42 patients with CAF were followed up at the three centres, with an overall rate of spontaneous closure of 17% (7/42 patients: 2/16 patients in Lille, 2/15 patients in Lyons, and 3/11 patients in Leuven).

Of the remaining 35 patients, 18 (51%) were closed by surgery or catheterisation (3/13 in Lyons, 8/14 in Lille, and 7/8 in Leuven).

It was decided to follow up the remaining 17 patients with echocardiography alone, either because of their age or because of the small size or asymptomatic nature of the fistulas.

Discussion

The main clinical manifestation of CAF is a continuous murmur over the abnormal communication site. The right coronary artery is most often involved in clinically detectable CAF (developing close to low pressure cavities). Fistulas most frequently involve the right ventricle, closely followed, in terms of incidence, by drainage into the right atrium or pulmonary artery. Left coronary fistulas are less common, but usually drain into the right ventricle or right atrium. Coronary fistulas rarely drain into the left atrium or left ventricle.

The incidence of this anomaly ranges from 0.2–0.4% in selected series of congenital heart disease patients, and its incidence in the overall population is estimated to be about 0.002%.^{15 16}

Congestive heart failure is the main complication of CAF and mostly occurs in the fourth decade or later, and rarely in neonates or during childhood, except if the QP:QS ratio is very high.¹⁷ Myocardial ischaemia, presumably resulting from the “coronary steal” syndrome, has been noted in 15–20% of cases.^{2 17} Atherosclerotic coronary artery disease, which occurs earlier and more frequently, is considered to be

Table 2 Cases of spontaneous closure or near closure published in the literature

Authors	Sex	Age at diagnosis	Murmur	Fistula course	QP:QS	Age at closure (years)
Morgan <i>et al</i> ²	Male	3.5 years	Continuous 4/6	LAD → RV	–	4.5
Jaffe <i>et al</i> ³	Female	29 years	–	RCA → RV	1.1	44
Mahoney <i>et al</i> ⁴	Female	6 months	Continuous 3/6	LCA/RCA → RV	1.4	5
Shubrooks <i>et al</i> ⁵	Male	10 years	S-D 4/6	LAD → RV/RA	1	15
Griffiths <i>et al</i> ⁶	Female	2 months	Continuous 3/6	LAD/RCA → RV	–	5.5
Hackett <i>et al</i> ⁷	Female	1 year	Continuous 4/6	RCA → RV	1.4	4
Mühler <i>et al</i> ⁸	Male	5 months	Continuous	LAD → RV	1.05	4
Muthusamy <i>et al</i> ⁹	Male	41 years	–	LAD → PA	1	42.5
Tomita <i>et al</i> ¹⁰	Female	1 year	Continuous 3/6	LAD → RV	1.4	4
Nakatani <i>et al</i> ¹¹	Male	55 years	–	RCA/LAD → PA	1	59
Farooki <i>et al</i> ¹²	–	10 months	–	RCA → RV	1.5	Between 8 and 10
–	–	2 months	–	RCA → RV	2.8	4
Jazra <i>et al</i> ¹³	Male	35 years	S-D 2/6	LAD → PA	–	36
Wong <i>et al</i> ¹⁴	Female	4 years	Continuous	LCA → RV	1.1	–
–	Female	4 months	Systolic	RCA → RV	1	–

S-D, systolo-diastolic murmur; see table 1 for key to other abbreviations.

one of the major potential complications of CAF.¹⁶ Bacterial endocarditis is unusual (3–4%) but can occur at any age.^{16–18} Progression into pulmonary hypertension or dilated hypokinetic myocardialopathy has been rarely reported.¹⁶

Elective closure by surgery or catheterisation can be recommended in adults, either on the basis of symptoms or on the grounds that complications will appear in the future and that operative mortality rate is low.^{15–17, 19} It is also indicated in symptomatic children, although two cases of spontaneous closure were reported in children who had initially presented with congestive heart failure.^{12, 19} In such cases, preoperative evaluation must accurately define CAF anatomy and physiopathology. It requires selective coronary angiography showing involvement of the coronary artery and collateral vessels, the CAF course, the receiving cardiac chamber, and the exact communication site. Only after all these investigations have been completed will the closure technique be chosen, either by catheterisation or surgery, with or without extracorporeal cardiopulmonary bypass. Embolisation by coils or balloons reduce hospitalisation time and avoid sternotomy and cardiopulmonary bypass.^{19, 20}

In asymptomatic children, systematic closure by surgery or embolisation is controversial. The percentage of spontaneous CAF closure needs to be verified, in consideration of its uncertain natural history and also because spontaneous closure is estimated to occur in 1% of all reported cases of CAF.¹² However, as most patients are asymptomatic, the continuous murmur is not always detected in children or infants, and consequently cases of spontaneous closure may remain undetected and the above rate is probably an underestimate.

These features (asymptomatic fistulas, undetected cases) probably explain why in the present series a closure rate of 17.5% was found, considerably higher than the usual 1%. The increasing use of echocardiography, adding to a possible recruitment bias, may have also contributed to this figure.

So far, 15 reports of spontaneous CAF closure, as observed by angiography or echocardiography, have been published (table 2). Closure was complete in 11 cases, incomplete in two cases, and clinically suspected in another

two cases on account of the disappearance of the murmur.

The mechanism responsible for closure of such fistulas is unknown. Jaffe and colleagues suggested that shear induced intimal damage resulting from shunt flow may cause atheromatous changes at the level of narrow fistulous communication with the heart.³

Literature data are very disparate, including children and adults, small and large shunts, and incomplete follow up. It is therefore difficult to define a clear management strategy for CAF. Nevertheless, it appears that spontaneous closure can be expected in infants under 2 years old if the CAF drains into the right heart, especially the right ventricle, with a QP:QS ratio below 1.5. In such an “ideal” case, closure can be observed between 1–6 years following diagnosis.

For such patients, it is therefore possible to wait, as was done in this series, before intervening. The risk of complications during this short period is very low. Conversely, CAF in adults, in patients with a large left-to-right shunt, in association with a complication, or in symptomatic patients probably need to be closed.

In most of the published data, spontaneous CAF closure was revealed at angiography. To our knowledge, this series is the first to describe patients whose follow up by colour Doppler echocardiography permitted monitoring of spontaneous CAF closure by non-invasive techniques. Thus, two dimensional Doppler mode echocardiography can help diagnose and evaluate the haemodynamic repercussions of CAF and permits non-invasive follow up if a “waiting attitude” is preferred.

In conclusion, the classical view that nearly all patients with CAF are potential candidates for surgical or percutaneous correction, in order to prevent significant and potentially fatal complications, needs to be reviewed. As the natural history of CAF remains unclear, asymptomatic patients, in particular children under 7 years old with small shunts to the right ventricle, can be periodically followed up by echocardiography rather than be subjected to operative closure, albeit by catheterisation.

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